## Board Review Aortic Valve, HCM, Systemic Disease

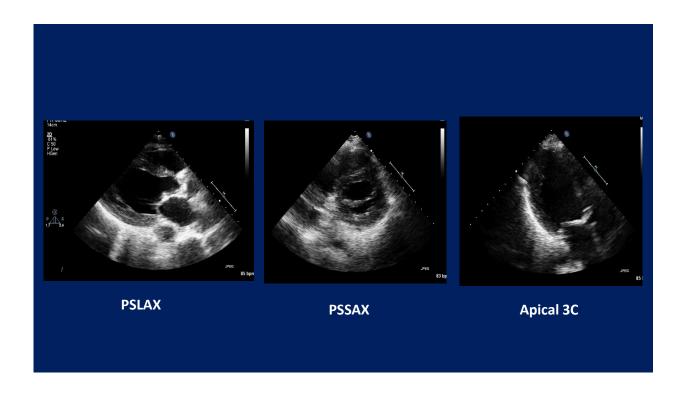
Dennis A. Tighe, MD, FASE
University of Massachusetts Medical School
Worcester, MA

# Which of the following statements regarding the obstructed/thrombosed prosthetic heart valve (PHV) is correct?

- 1. PHT ≤ 130 msec is the single best indicator of prosthetic mitral obstruction.
- 2. Taking heart rate into account is not necessary when assessing transmitral gradients.
- 3. Pannus in-growth is more common with PHVs in the aortic position than in the mitral position .\*\*
- 4. A peak velocity ≥2.5 m/sec suggests significant aortic PHV stenosis.
- 5. Randomized, controlled trials have demonstrated that bolus infusion of rt-PA is the fibrinolytic regimen of choice.

#### Case 1

- A 54 year-old woman with hypothyroidism presents to her PCP with worsening shortness of breath.
  - Systolic and diastolic murmurs are auscultated.
  - Transthoracic echocardiography is requested for further evaluation.



Q1. Echocardiography confirms the presence of aortic stenosis (orifice area 0.6 cm²) and identifies the presence of moderate aortic regurgitation. Mitral valve thickening is also observed. Which of the following constitutes the most likely etiology for the valvular abnormalities:

- 1. Age-related degenerative valve disease
- 2. Rheumatic heart disease
- 3. Annular calcific disease
- 4. Carcinoid heart disease
- 5. Radiation-associated valve disease\*\*

## Q2. Which of the following conditions could be an expected complication resulting from the disease process causing these left-sided valvular abnormalities?

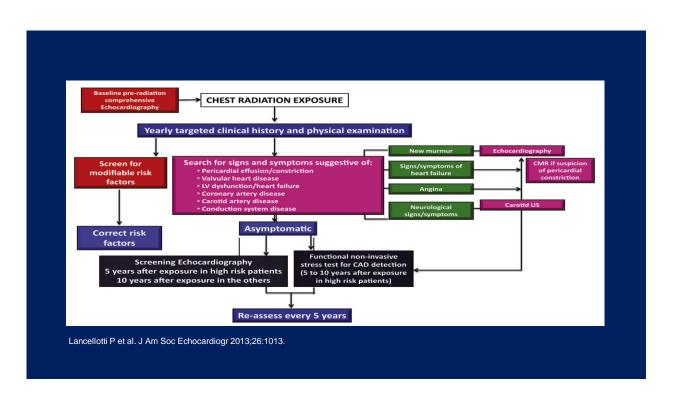
- 1. Flushing
- 2. Constrictive pericarditis\*\*
- 3. Coronary artery spasm
- 4. Hypertrophic cardiomyopthy
- 5. Cardioembolic stroke

#### **Radiation-Associated Valve Disease**

- Frequent complication
- Regurgitant lesions > Stenotic lesions
  - Left sided > right sided
- Risk greater with ≥30 Gy
- Women > men
- Suggestive echocardiographic appearance
  - Calcification and thickening of aortic-mitral curtain
  - Anterior changes more profound than posterior (vs MAC)
  - No leaflet doming/commissural involvement (vs RHD)
  - Aortic root calcification increases the likelihood
- Progressive
- Periodic screening required

### **Radiation Therapy**

- Cardiovascular complications
  - Coronary artery disease
  - Cardiomyopathy
    - Restrictive or dilated
  - Pericardial effusion
  - Pericardial thickening/constrictive pericarditis
  - Conduction system/arrhythmias
  - Valvular heart disease
  - Carotid artery disease

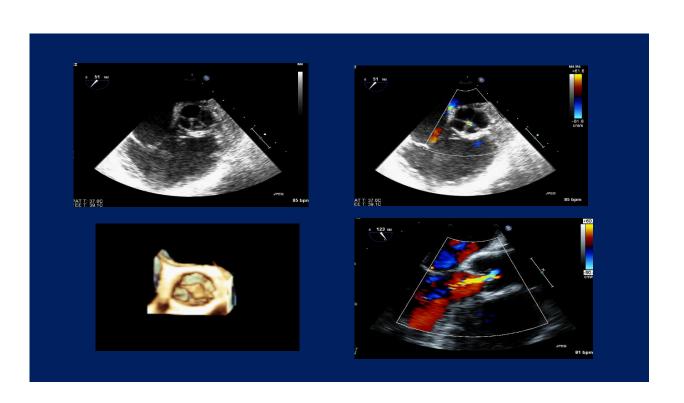


## Which of the following statements concerning prosthetic heart valve regurgitation is correct?

- A. Pseudo-regurgitation is an issue most often encountered during performance of TEE.
- B. Any degree of regurgitation indicates dysfunction of a mechanical heart valve.
- C. Structural valve deterioration is an uncommon cause of pathological regurgitation.
- D. A mitral bioprosthesis is less likely to suffer structural valve deterioration than is an aortic bioprosthesis.
- E. Annular dehiscence is most often a consequence of infective endocarditis.\*\*

Case 2

- A 56-year old woman presents with shortness of breath. Right heart volume overload due to ostium secundum ASD is found on echocardiography. She is referred for percutaneous ASD closure. RHC revealed PASP 29 mm Hg and PVR <2 WU. Non-obstructive CAD was found.
- TEE performed at the time of the procedure confirms the presence of the ASD and documents adequate surrounding rims and appropriate PV drainage. An incidental finding is made.



## Q1. Which of the following courses of action should you recommend?

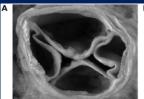
- 1. Inform your interventional colleague that the procedure should be ceased immediately and discuss the findings with the patient and her family.
- 2. Obtain an urgent CT surgery consult.
- 3. Provide additional antibiotic coverage for oral cavity organisms given a high-risk of infective endocarditis.
- 4. Continue with the planned procedure and discuss the incidental finding and its implications with the patient and referring cardiologist.\*\*

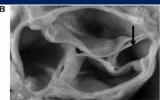
## Q2. Which of the following statements concerning this valvular abnormality is correct?

- A. Aortic stenosis is the predominant hemodynamic abnormality.
- B. Aortic regurgitation is the predominant hemodynamic abnormality.\*\*
- C. Long-term survival with this condition is poor.
- D. Aortic dilatation is not commonly found in association with this lesion.
- E. Aortic dissection is strongly associated with this valvular abnormality.

#### **Quadricuspid Aortic Valve**

- Rare congenital abnormality (<0.05% frequency)</li>
  - Recent single-center review showed frequency of 0.006% of all echocardiograms
- Hurwitz and Roberts classification
  - Types A and B most frequent





Tsang MYC et al. Circulation 2016;133:312-319.

### **Aortic Valve Findings**

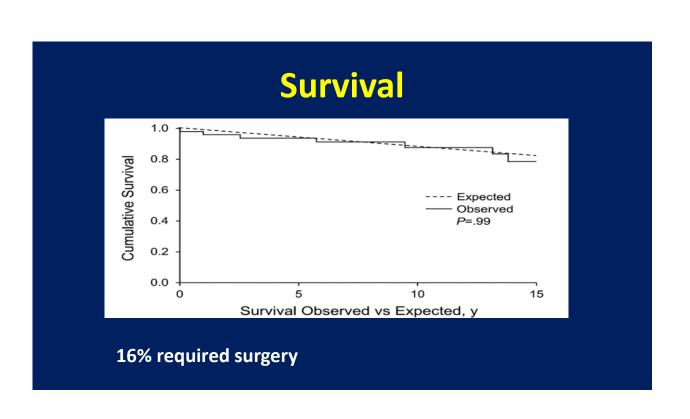
QAV Subtype*	Functionally Normal AV,† n	AR Only (With Moderate or Greater AR), n	Aortic Stenosis Only, n	Both AR and Stenosis, n
A (n=16)	0	16 (6)	0	0
B (n=16)	2	12 (3)	0	2
C (n=8)	2	5 (2)	1	0
D (n=4)	0	3 (0)	0	1
E (n=1)	0	1 (0)	0	0
F (n=4)	0	4 (2)	0	0

Aortic regurgitation is the predominant hemodynamic abnormality

Aortic stenosis present in only 8%

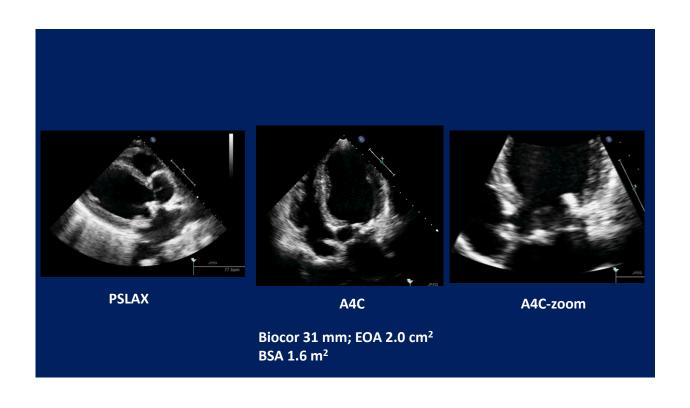
### **Associated Findings**

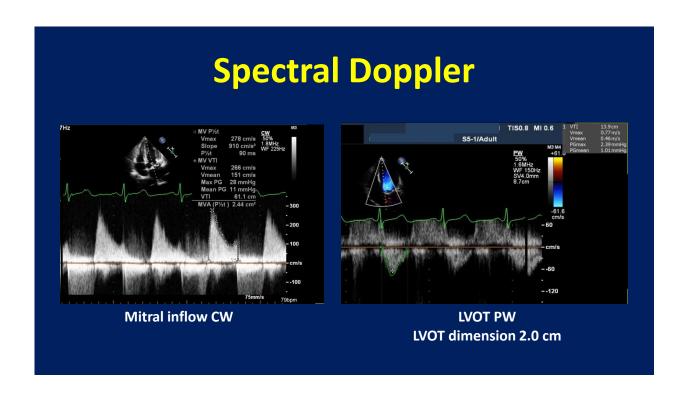
- Aorta
  - Dilatation 29%
    - · Usually mild
- Other cardiac lesions
  - MVP/bowing
  - TVP
  - Pulmonary valve stenosis
  - ASD
  - VSD
  - Coronary anomaly



#### Case 3

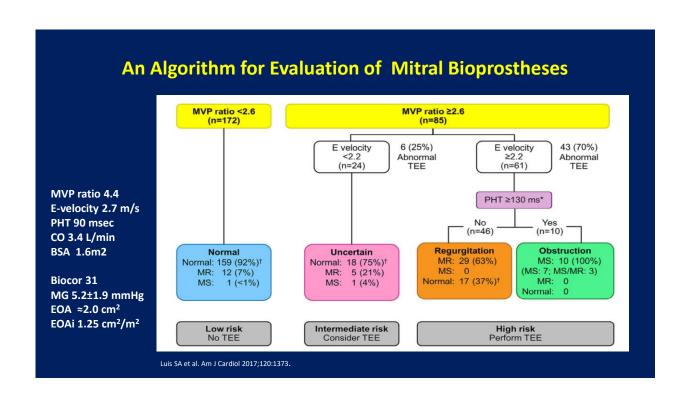
- A 60 year old man with prior MSSA infective endocarditis s/p MVR (31 mm Biocor valve) in 2008 and cirrhosis presents with shortness of breath.
- A heart murmur is heard.
- An echocardiogram is requested.

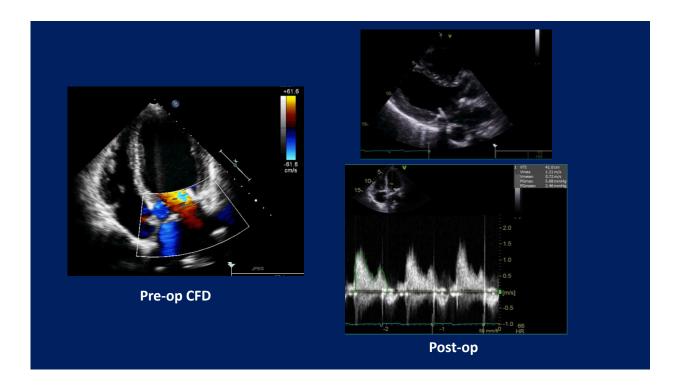




## Which of the following statements best characterizes the function and further evaluation of this PHV?

- 1. Significant regurgitation is likely present. TEE should be performed to confirm.\*\*
- 2. Obstruction is likely present. TEE should be performed to confirm.
- 3. Patient-prosthesis mismatch is likely present. TEE is not required to confirm prosthetic function.
- 4. A high cardiac output state is present. TEE is required to confirm normal prosthetic function.
- 5. The prosthesis is functioning normally. No further assessment is needed.



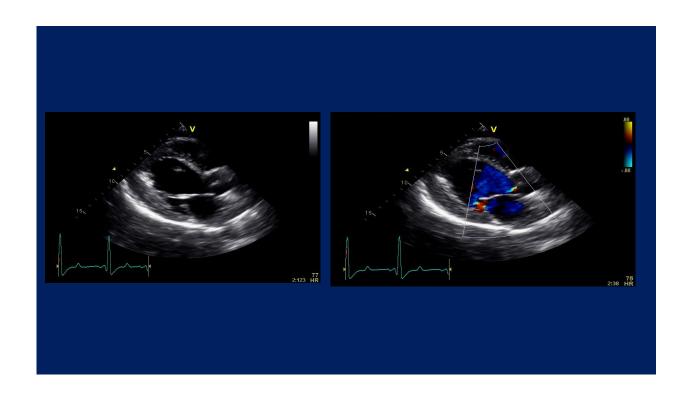


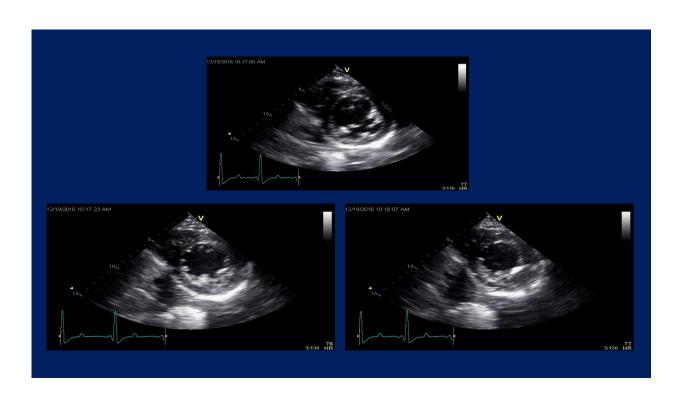
#### **Choice Explanations**

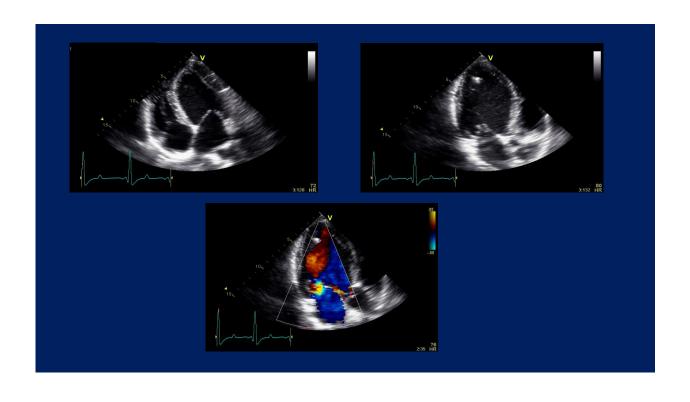
- 1. Significant regurgitation is likely present. TEE should be performed to confirm is CORRECT.
  - Observe that LV fct is hyperdynamic while the HR is in normal range (79/min). The CO is only 3.4 l/min. The DVI is markedly increased, the E-wave velocity is increased and the PHT is only 90 msec. The calculated EOAi was 1.25 cm²/m². This data suggests that prosthetic dysfunction due to significant regurgitation is likely present. TEE is recommended to confirm this suspicion.
- 2. Note that the PHT was 90 msec—this makes PHV dysfunction due to obstruction unlikely.
- 3. PPM is characterized by high trans-prosthetic valve gradients with normal prosthetic function. The high value of the PHV-DVI makes normal PHV function unlikely and the EOAi is not <1.2 cm<sup>2</sup>/m<sup>2</sup>.
- 4. While the 2D images of the LV show hyperdynamic function (due to severe MR in this case), the calculated CO was only 3.4 I/min which is not consistent with a high-CO state.
- 5. The overall picture with a new murmur, hyperdynamic LV function, and abnormal Doppler echocardiography of the PHV strongly suggests that prosthetic dysfunction is present and normal PHV function thus is unlikely.

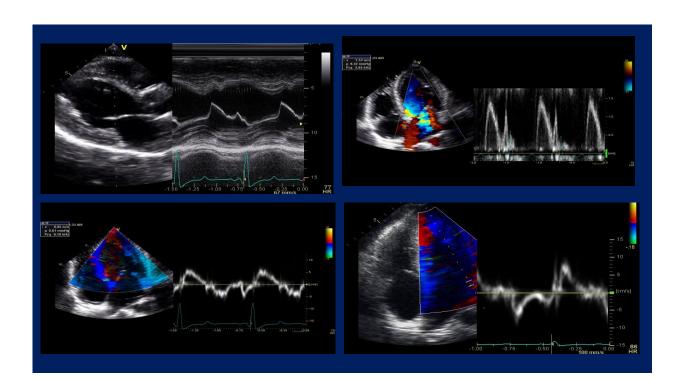
#### Case 4

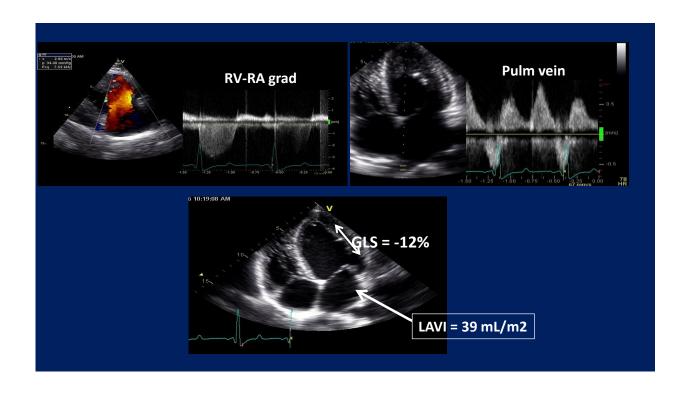
- Echo number 25 of a long day.
- A 22 year-old man previously cared for in pediatric cardiology clinics is referred by his new cardiologist for echocardiography to evaluate mitral regurgitation and LV function.
- He has seen multiple subspecialists over the years. The patient has ESRD and was recently initiated on HD. He has poorly-controlled HTN and his CBC/diff is distinctly abnormal.

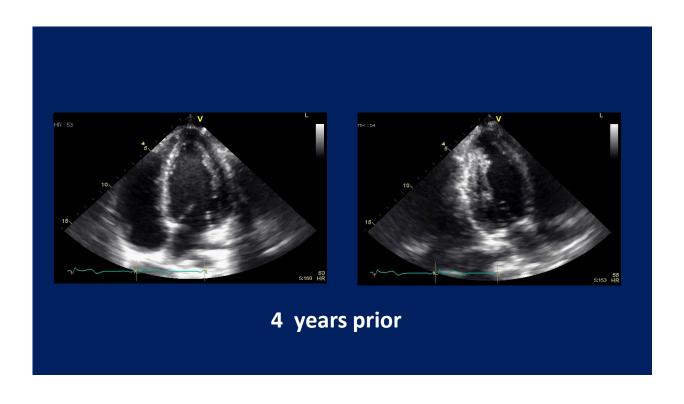












## Q1. Based on the available data, how would you best characterize his left heart function?

- A. Normal LVEF, elevated mean LAP, elevated LVeDP\*\*
- B. Normal LVEF, elevated mean LAP, normal LVeDP
- C. Normal systolic function with normal filling pressures.
- D. Normal systolic function with elevated filling pressures.

## Q2. Which of the following conditions is the most likely etiology for the observed echo findings?

- A. Amyloidosis
- B. Apical hypertrophic cardiomyopathy
- C. Hypertensive heart disease
- D. Hypereosinophilic syndrome\*\*
- E. Rheumatic heart disease

#### **Lab Studies**

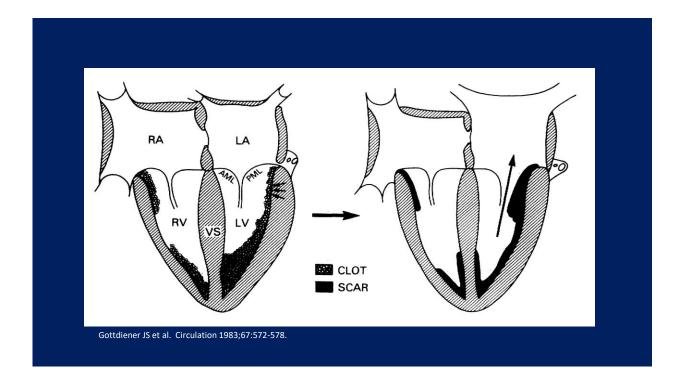
- BMP
  - Na 139; K 3.7; Cl 100; Co2 29; BUN 15; Cr 6.4;eGFR 11.
- CBC/diff
  - WBC 12.7; Hgb 8.5; Hct 25.7.

Mankad R et al. Heart 2016;102:100-106.

- PMN 18%; Lymph 13%; Eos 64%; Baso 2%.

### **Hypereosinophilic Syndome**

Table 3 Stages of cardiac pathology in HES				
Stages of cardiac involvement in HES	Characterisation of stage			
Acute necrosis	Eosinophilic myocarditis with eosinophilic and lymphocytic infiltration. Myocardiac necrosis and apoptosis with rare microembolic phenomena. Typically no other cardiac symptoms. Rarely can exhibit a fulminant course.			
Thrombotic stage	Thrombus along damaged endocardium. Thrombi within apices of one of both ventricles, and can encroach on base of the heart into the subvalvular regions. Embolic phenomena can occur.			
Fibrotic stage	Thrombi are replaced by fibrosis. Scarring occurs. Restrictive cardiomyopathy with signs and symptoms of left or right sided heart failure. Fibrosis that occurs at the base of the heart can lead to valvular regurgitation.			
HES, hypereosinophilic syndrome.				



As recommended by the 2014 AHA/ACC Valvular Heart Disease Guideline, which of the following statements regarding follow-up of prosthetic heart valves by echocardiography is true?

- A. Annual TTE is reasonable staring at 5 years following mechanical valve replacement.
- B. An initial TEE should be performed routinely to assess valve hemodynamics within 2 months of implantation.
- C. Change in clinical status should prompt early echocardiography.\*\*
- D. Annual TTE is reasonable starting at 5 years following bioprosthetic valve replacement.

